

becoming incontinent of urine, how would we be judged?

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Panayiotopoulos syndrome

A common benign but underdiagnosed and unexplored early childhood seizure syndrome

Epilepsy affects 1% of the general population and 4% of children, encompassing heterogeneous seizure syndromes.¹ These are defined by distinct aetiology, age at onset, seizure type, and electroencephalographic features, which taken together provide the key to diagnosis, prognosis, and optimal management. Over the past two decades various distinct paediatric epilepsy syndromes, such as rolandic epilepsy, have been formally recognised.² Panayiotopoulos syndrome is a new idiopathic childhood epilepsy, recently recognised by the International League Against Epilepsy.^{2,3} It is common, benign, and may mimic other common illnesses.

Awareness of this syndrome is important for all professionals who care for children with epileptic seizures, including general practitioners and community nurses, paediatricians and paediatric neurologists and clinical neurophysiologists, for the following reasons. Firstly, it is common. It probably affects about 13% of children of 3-6 years old with one or more non-febrile seizures (peak age 4-5 years), and 6% of the age group 1-15.^{4,5} Secondly, seizures can be prolonged, may mimic non-epileptic disorders, and may vary in severity from trivial to apparently life threatening—implying that the diagnosis may need to be considered in a variety of clinical settings and by medical professionals of different specialties. Thirdly, it is benign—its recognition therefore can provide firm reassurance to families in situations that can be very alarming. Finally, clinical research, necessary in any new syndrome, would require a multidisciplinary approach.

Panayiotopoulos syndrome can be best defined as idiopathic susceptibility to early onset benign childhood seizures with electroencephalographic occipital or extra occipital spikes, and manifests mainly with autonomic seizures.⁴ Large independent studies have accumulated impressively concordant information on the clinical and electroencephalographic features of this syndrome.⁴⁻⁹

Rolandic epilepsy, another common syndrome, affects 15% of children with seizures and has as good a prognosis as febrile convulsions. Seizures usually start between 7-9 years of age, occur mainly during sleep and consist of hemifacial convulsions, speech arrest, oropharyngolaryngeal movements, and hypersalivation. EEG shows centroparietal spikes. Gastaut's occipital epilepsy is much less frequent, manifests with mainly

visual seizures, and has less predictable outcome. Electroencephalography shows occipital spikes.²

Autonomic seizures are the hallmark of the Panayiotopoulos syndrome.⁴⁻⁹ Autonomic symptoms and signs (mainly vomiting) occur from the onset in 80% of seizures, with half of them lasting for more than 30 minutes to hours, thus amounting to autonomic status epilepticus. Two thirds of the seizures occur during nocturnal sleep or brief daytime naps. In a typical daytime seizure the child looks pale, complains, "I want to be sick," and vomits. If in sleep, the child wakes up with similar complaints or is found vomiting, confused, or unresponsive. Vomiting occurs in three quarters of seizures. Other autonomic manifestations may occur either concurrently with vomiting or later in the course of the seizure, and include pallor, mydriasis, cardiorespiratory, gastrointestinal and thermoregulatory alterations, incontinence, and hypersalivation. In at least a fifth of the seizures the child becomes unresponsive, pale, and flaccid (ictal syncope)⁴ either before convulsing or in isolation.

Behavioural disturbances, headache, or various non-painful cephalic sensations are common particularly at onset. More conventional manifestations of seizures often ensue: the child becomes confused or unresponsive, eyes may deviate to one side (in 60%) or the patient may stare. Half of the seizures end with hemi or generalised convulsions. Other, less frequent ictal features include speech arrest, hemifacial spasms, visual hallucinations and oropharyngolaryngeal movements, suggesting a maturation related continuum with Rolandic epilepsy.¹⁰

Diagnosis of Panayiotopoulos syndrome may be easily missed—mild and brief ictal autonomic symptoms in the presence of clear consciousness would suggest trivial non-epileptic conditions such as atypical migraine, gastroenteritis or syncope, while prolonged and severe attacks may simulate life threatening insults such as encephalitis, for which many of these children are treated.^{3,4,9}

Characteristically, even after the most severe seizures and status, the child is normal after a few hours of sleep—this is both reassuring and diagnostic. Electroencephalography, which should be done after a first non-febrile seizure, is confirmatory. This usually shows multifocal spikes at various locations.^{3-5,7,11}

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Though occipital spikes predominate they are neither a prerequisite nor specific.¹² Normal recordings may occur in 25% of children¹¹ and should prompt an EEG during sleep to activate the spikes; on strong clinical suspicion a sleep EEG should be done. A useful rule of thumb is that Panayiotopoulos syndrome should be considered if a normal child with a single or a few seizures has an EEG with multifocal spikes.^{3 4 10 11}

Panayiotopoulos syndrome is remarkably benign. Remission usually occurs within two years from onset.^{4 6-9} A third of these children have a single seizure, and only 5-10% have more than 10 seizures that may be very frequent sometimes but the outcome is still favourable.⁴ Lengthy seizures do not appear to result in residual deficits or have adverse prognostic significance. One fifth of children with this syndrome may develop other types of infrequent, usually rolandic seizures, but these also remit before the age of 16 years.

It follows that treatment with antiepileptic drugs (mainly carbamazepine) is usually unnecessary but may be considered in children with multiple seizures. The decision should also take into account a likely traumatising parental experience as in febrile convulsions.^{3 4 8} Appropriate advice by the family doctor is expected to shape parental attitude and prevent chronic anxiety.

There is a lot more to learn about this syndrome. Prospective epidemiological and clinical studies should assess the actual prevalence, delineate the clinical spectrum, and define possible clinical and electroencephalography markers of atypical clinical presentations and complicated evolution that is occasionally seen.⁴ Possible genetic links with the rolandic phenotype may provide further information about the age related continuum of "benign childhood seizure susceptibility syndrome."¹⁰

Finally, in the light of this syndrome, the concept of ictal syncope in childhood clearly needs to be evaluated

again. It is currently hypothesised that in Panayiotopoulos syndrome an inherent autonomic instability responds by generating autonomic seizures and status when cortical hyperexcitability triggers susceptible brain circuits.⁴ As the current epidemiological data seem to indicate, this hyperexcitable loop is mainly related to the early childhood and is short lived. Careful prospective and controlled studies of autonomic function will clarify the roles of the cortex and the brainstem in the generation and expression of seizures.

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Regulating cosmetic surgery

Members of the public would be better protected if they consulted their general practitioners first

Cosmetic surgery has become a growth industry and a public obsession. The demand for the top three procedures in the United States grew by 26% between 1999 and 2000, and this growth is mirrored in the United Kingdom.¹ The public perception of cosmetic surgery is that it is quick and easy. In fact most cosmetic surgery operations are extremely complex and require a high degree of anatomical knowledge and surgical skill as well as aesthetic appreciation.

The public's increasing interest is accompanied by a reduction in the provision of cosmetic surgery in the NHS, so that patients look to the private sector, financing their treatment through bank loans and finance agreements.^{2 3} These patients have been prey to organisations that offer discounts, privacy, and no waiting time but are not staffed by accredited surgeons.

Many patients do not seek a referral from their general practitioner because they fear an unsympathetic response or they feel that cosmetic surgery is not

fundamentally medical. Self referral to a clinic is an easier option.

Standards in cosmetic clinics vary, but the clinics often send a representative to the home of the patient in response to a reply to an advertisement. These representatives are not medically qualified but recommend operations and book dates for surgery, often offering discounts if the patient signs immediately. Starved of food before having general anaesthesia and having paid the fee, the patient briefly meets the surgeon before the operation.

New government regulations insist on preoperative consultations by the surgeon and ban surgery within two weeks of consultation.⁴ The regulations also insist that clinics are inspected regularly and that written information is realistic.

The training of junior surgeons in cosmetic surgery is proving an extremely contentious issue since less training is now provided in the NHS than ever before and in any case many cosmetic procedures were never